

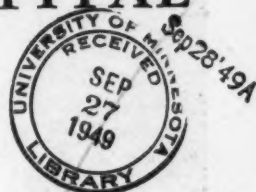
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of the

CHILDREN'S HOSPITAL

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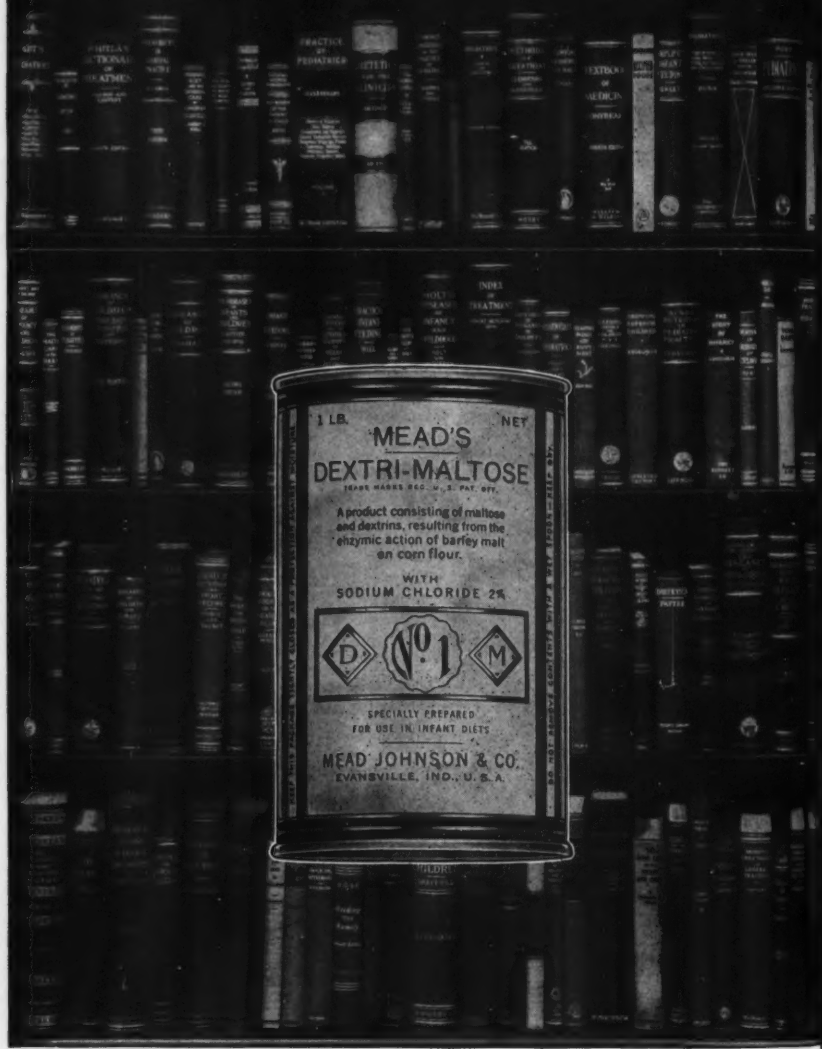
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VOLUME V

NUMBER 10



BACKGROUND



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CLINICAL PROCEEDINGS

OF THE CHILDREN'S HOSPITAL

13th and W Streets, Washington 9, D. C.

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SPECIAL REPORT

EARLY DIAGNOSIS IN CONGENITAL CAUSES OF CRIPPLING

Morris Tandeta, M.D.

INTRODUCTION

This paper represents a three months' survey of the clinical material seen in the Orthopedic Out-Patient Department at Children's Hospital, dealing specifically with the prevention of deformities that result from congenital malformations of the lower extremity. Both pediatrician and orthopedist have here a mutual interest with a common cause; namely, healthy, happy children unmarked by preventable physical stigmata. The pediatrician who is aware of a congenital abnormality of the lower extremity at the time of the routine newborn or regular monthly health examination can refer the case immediately to the orthopedist for evaluation, x-ray, and follow-up. The earlier the diagnosis is made, the sooner corrective treatment can be instituted, and the better the ultimate outlook. To this end the most common disabling malformations of the lower extremity, congenital dislocation of the hip and clubfoot, are considered in some detail. It is of historic interest that in the fifth century B.C. Hippocrates described congenital hip dislocation and clubfoot, outlining satisfactory management of both these entities. Yet the modern concept of early diagnosis and early conservative treatment, essentially the same as his, was not accepted by the profession until the turn of the last century.

Several hypotheses have been advanced to account for the occurrence of congenital anomalies. Two of the more widely accepted are the genetic and mechanical theories. The genetic theory explains congenital abnormalities as inherited mutations resulting from a defect in the germ plasm. The frequency with which a family history of congenital malformations is obtained, and the coincidence of other anomalies in the same individual tend to support it. Yet we are unable to correlate all our findings with the hypothesis of a primary germ cell defect. A corollary to this is the theory of early embryonic arrest, that is, cessation or interruption of normal development in the embryonic stage. This is hardly plausible because study of very early specimens has failed to disclose any such normal stage in development as is represented by the findings at birth. The mechanical theory, on the other hand, postulates that confinement of the fetus in utero makes him particularly susceptible to increased stress or strain, especially if for some reason the protective fluid envelopes should become deficient. Excessive compression of the fetal structures would then lead to stretching of soft

parts, relaxation of joints, or even direct trauma to the relatively unprotected extremities. Although this theory seems to explain why some deformities conform to shapes consistent with uterine molding, it does not take into account frequent changes in position of the fetus, nor is there evidence of intrinsic destruction, inflammation, or repair in any of these malformations. Sex distribution would also militate against a purely mechanical etiology. The associated neurogenic theory presupposes interference with the normal nerve supply to certain muscles, bringing about imbalance and overaction of antagonistic muscle groups. Observed clinical abnormalities can hardly be explained by exaggerated muscle pull alone.

From the experimental work of Warkany and others it is conceivable that hitherto unrecognized factors, such as irradiation, interference with fetal nutrition through faulty implantation of the ovum, placental disease, obstruction of umbilical blood flow, toxins, and inadequate maternal diet may also cause deformities. Only by the correlation and integration of many isolated facts from various sources will an adequate interpretation of the origins of congenital anomalies be forthcoming.

CONGENITAL DISLOCATION OF THE HIP

Congenital dislocation of the hip is a misnomer, because rarely is there evidence of true hip displacement at birth. A better term would be congenital dysplasia of the acetabulum predisposing to dislocation of the femoral head. In the newborn, the concave surface of the acetabulum is made up of three mounds representing the epiphyses of the ilium, pubis, and ischium, the so-called tri-radiate cartilage, and articulating with it is the cartilaginous anlage of the head of the femur.

Congenital dislocation occurs more often in the hip than in all other joints combined. It is quite prevalent in the Mediterranean countries, especially certain sections of Italy, where the incidence is as high as three per thousand births, but is relatively rare in this country. It is more common in females in a ratio of six to one and about 30-40% are bilateral, with the left side predominating in unilateral cases. In at least 20% a familial history of congenital hip is obtainable. Only about one of four dysplastic hips, however, progresses to the stage of true dislocation. Since this affection is limited to the human species and is more common in females, the factors of erect posture with widened pelvis and more perpendicular innominate bone must be assumed contributory to dislocation.

The dysplastic hip shows a poorly developed acetabular roof, an abnormal obliquity of its rim, and disproportion in the size of the socket and femoral head. Any gradation of this condition may exist, from slight obliquity of the rim to almost complete obliteration and flattening of the acetabulum. The earlier the onset of dysplasia in embryonic life, the

greater the degree of resulting incongruity of the joint. Scaglietti is of the opinion that congenital hip dysplasia is always bilateral, and that unilateral cases represent spontaneous healing of the unaffected side.

In Europe the difference between sub-luxation of the hip and true dislocation is stressed. Opinion in this country has it that sub-luxation or pre-luxation merely represents an early stage in the development of actual displacement of the femoral head. By sub-luxation is meant a slight lateral displacement of the head of the femur from its socket without any upward progression or loss of contact with it. True dislocation appears later upon the assumption of the erect posture. Weight bearing with attendant muscular contraction pulls the head upward and backward on the side of the iliac crest, with concomitant anteversion or twisting of the femoral neck. This displacement is accompanied by stretching and twisting of the joint capsule, which becomes constricted at its mid-portion in the process. Following dislocation of the head, which acts as a stimulus for normal development of the acetabular cavity, the already shallow acetabulum fills with connective tissue.

In relation to this problem, Kleinberg and Lieberman x-rayed a large number of pelves of infants, and worked out criteria for detection of abnormal hips. They determined the angle of inclination of the iliac segment of the acetabulum with the horizontal Y-line, a line drawn through the tips of the ilia and the centers of the tri-radiate cartilages. This angle, designated the acetabular index, averages 27.5° in the newborn and 20° at two years of age. In congenital dysplasia of the hip the angle is increased, giving an acetabular index of 30 or greater. Needless to say, uniformity in x-ray technique is of utmost importance for accurate, dependable measurements.

Diagnosis of congenital hip dysplasia can be made in early infancy. Poli, of Milan, made a diagnosis at birth in 6.3% of 11,532 hips, and added 4.1% in the first year. Bost, et al., reported 15% of 72 hips diagnosed under one year, 52% under two years, and 33% under three years. The youngest age at diagnosis in this group was eight days. The earliest evidence of abnormality is usually limitation of abduction of the affected hip as the infant lies on its back with knees and hips flexed. By the second month inspection reveals asymmetrical creases on the medial sides of the thighs. The gluteal and inguinal folds also appear to be higher on the involved side. At the age of three months, the affected leg is turned outward, and the infant does not seem to move it as freely as the other one. If the infant is placed on its back with hips and knees flexed and feet touching the table, the knees appear to be at different levels due to apparent shortening. Later on there is a tendency to hold the hip in slight flexion, and any attempt at extension meets with resistance. Alternate pulling and pushing on the ex-

tremity may give a sensation of joint instability described as "telescoping." Because of the lateral prominence of the greater trochanter, the affected buttock takes on the rounded adult female contour. In bilateral cases, there may be a detectable widening of the perineum.

Any of these signs appearing before the weight-bearing age should suggest the possibility of hip dysplasia. After the infant begins to walk, the mother usually makes the diagnosis. The limping and turning outward of the leg are characteristic of dislocation while in bilateral involvement there is a peculiar waddling gait with exaggerated lumbar lordosis.

X-ray examination of the pelvis is mandatory at the slightest suspicion of a congenital hip. Early positive findings include increased obliquity of the acetabular roof, shallow socket, anteversion of the femoral neck, and lag in development of the capital epiphysis. In late cases, there is a subluxation or actual dislocation of the femoral head with a markedly abnormal acetabulum.

Principles of management have evolved gradually from the cumulative experiences of many able men in orthopedics. Among these the names of Lorenz and Putti stand out.

Treatment has for its aim the restoration of a good anatomical relationship between the head of the femur and its socket in order to provide a stimulus for normal development and function. The method of choice is that outlined by Vittorio Putti, and consists of fixation of both lower extremities in wide abduction and internal rotation. Some slight variations in his method of immobilization may be utilized, but the principle is now universally accepted. Treatment started in early infancy offers the best prognosis for a normal joint, because at this time the tissues are soft and supple, secondary bone changes are minimal, and the innate power of growth is able to overcome the slight malformation. Scaglietti reported 94% satisfactory results in a series of 478 dysplastic hips when treatment was instituted in the very first year. Bost, et al., got 80% satisfactory reductions in 72 hips treated before the age of two. Gill reported only 55% satisfactory hips of 22 treated in infancy. After the age of two or three the prognosis for reduction of a dislocated hip is poor. In a series of 456 hips Severin obtained satisfactory reduction in only 7% with a re-dislocation rate of 15% when treatment was instituted after the age of two years.

CONGENITAL CLUBFOOT

Congenital clubfoot or talipes deformity is a collective term referring to an abnormality of the foot and ankle present from birth. The type of deformity is best described with reference to the position of the foot in the vertical and horizontal planes. The terminology is derived from the

Latin "pes" meaning foot and "talus" the ankle. "Equinus" refers to plantar flexion at the ankle, since the deformity causes toe walking from the word for horse, while "calcaneus" is just the opposite with prominence of the heel. "Varus" means turning inward as opposed to "valgus", or turning away from the mid-line. Talipes equinovarus, in which there is adduction of the forefoot and inversion of the foot with plantar flexion at the ankle, occurs about once in a thousand births, constituting 75% of clubfoot deformities. Inversion or supination of the foot is due to the inward rotation of the os calcis under the talus. The other types, talipes equinovaglus, in which the foot is everted, calcaneovarus and calcaneovalgus with dorsiflexion of the foot and prominent heel, represent the remaining 25% of clubfeet. The following description will be limited for the most part to talipes equinovarus, which is the most persistent and difficult to correct, but the principles of management are essentially the same for this entire group.

Clubfoot is more common in males in a ratio of six to four, and from 40 to 50% are bilateral. Associated anomalies are quite common. A familial history can be obtained in about 20% of cases.

Although it is generally believed that no one etiological factor is responsible for clubfoot, the mechanistic theory has an able and staunch supporter in the English surgeon, Denis Browne. He is of the opinion that pressure applied to the soles of the feet in utero results in calcaneous, and compression of the outer borders of the feet as they swing in causes equinovarus. The degree of deformity then depends on the amount of force applied, so that in mild clubfoot deformities, such as metatarsus varus, only the forefoot meets the compressive force, while in talipes equinovarus the whole foot is molded to conform more or less to the uterine curvature. Brockman believes that the deformity consists primarily of a dislocation of the head of the talus, much as the femoral head in the congenital hip.

Diagnosis of clubfoot is possible immediately after birth. In talipes equinovarus the foot is in adduction, inversion, and plantar flexion. The skin is usually stretched thin and dimpled on the outer aspect with creasing apparent along the sole and inner border. Scarring from pressure may be evident on the dorsal surface. The lateral malleolus may appear unduly prominent in contrast to the poorly developed medial one. Due to marked muscle imbalance caused by overstretching of the extensors and peroneals and contracture of the medial group of muscles, the deformity is quite fixed. Correction does not follow gentle stroking or stretching, as one would expect of the normally elastic foot.

X-ray examination reveals the presence of the ossification centers for the cuboid, calcaneus, and talus, with the latter overriding the calcaneus, and fixation of the foot in the characteristic position.

Treatment of clubfoot may be instituted as soon as the newborn regains his birth weight and accustoms himself to his new environment, usually about the second week. The aim of treatment is correction of deformity and restoration of normal function. This latter goal is the more difficult of accomplishment. The sooner treatment is started, the more responsive the structures are to manipulation, the more the growth factor can be utilized in obtaining correction, and therefore the more perfect the end result. The principle involved is to reverse the deformity completely and to maintain the foot in its new position until muscle function is adequate to prevent recurrence. Gentleness is of paramount importance in preventing soft tissue damage with resultant hemorrhage, scarring, and decreased flexibility at the joints.

In this country orthopedists favor the Kite method of treatment, which is essentially that of repeatedly modified or wedged casts. When treatment is begun early, this method gives excellent results. The Denis Browne splint is preferred, however, in the British Commonwealth of nations. It has the advantage of wide adaptability in all types of talipes deformity, and it utilizes the kicking of the infant to strengthen the involved muscles. This splint is made up of individual foot-pieces which are fixed to a crossbar of aluminum. The feet can be maintained in almost any position desired by proper adjustment of the pieces or bending of the malleable crossbar. Other types of fixation have been proposed such as rubber band traction and a cohesive bandage, but these have not withstood the test of time. All of these methods require constant care and cooperation of the parents, as well as periodic checking by the surgeon. Ideally the surgeon strives for over-correction or reversal of the deformity in stages, the adduction first, then inversion, and finally the equinus. This is important for securing excellent function and normal gait. Treatment may be judged satisfactory only when the forefoot points outward at an angle of about 20° , and there is a full range of motion in the tarsal joints.

Since corrective management of these conditions is entirely within the province of the orthopedic surgeon, it is necessarily outside the scope of this short presentation. However, too much emphasis cannot be placed on the early recognition of congenital deformities by the pediatrician. Lack of interest by the latter may necessitate severe treatment of many years duration in cases in which only mild therapy of a few months is otherwise possible. The pediatrician is the only one in a position to initiate remedial action by making the diagnosis and advising the family of the necessity for orthopedic consultation.

In order to give some idea of the prevalence of congenital abnormality of the lower limb, use is made of the out-patient records of this hospital. During the three month period from December 1948 through February

1949 there were 19,833 out-patient visits to this hospital. In the orthopedic clinic there were 386 visits for the same period of time, representing 2% of the total. Of this number 78 patients were 2 years of age or under. In 63 of the infant age group there was involvement of one or both lower extremities, while only 8 had miscellaneous and upper extremity complaints. There were 2 cases of fractured clavicle, and only 5 patients showed no evidence of abnormality. Colored infants constituted 70% of patients.

Of the 63 infants with involvement of the lower extremity, 38 or 60% were due to congenital abnormality. The remaining, including 4 cases of external rotation of one or both legs due to the habit of sleeping in the prone position, were mostly postural deformities. Eight showed evidence of healed rickets. No fractures were seen in this group. There were 8 cases of talipes equinovarus, 20 related deformities of a lesser degree, and 5 of the calcaneovalgus variety. Only 2 cases of congenital hip were diagnosed.

These figures have no actual statistical significance because they represent a very limited study in time as well as subject matter. The patients too are selected rather arbitrarily, since only those deemed needy of free care are seen at the clinic. Many years of careful patient follow-ups would be required for completion of a study that would allow valid conclusions. In broad general terms at least we can deduce that congenital deformities are much more with us than many medical entities of which are we ever mindful.

SUMMARY

1. Disability and physical stigmata due to congenital dislocation of the hip and clubfoot are preventable.
2. Cognizance of the presence of these anomalies in early infancy leads to earlier diagnosis.
3. Early diagnosis and early treatment offer the best prognosis for correction of these deformities.

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OCULAR TORTICOLLIS

Case Report No. 158

Worth Sprunt, M.D.

G. W., a 5½ year old white male, was first seen in the Children's Hospital Orthopedic Clinic in February 1949 with a chief complaint of tilting of the head to the left.

He was a full-term baby and perfectly normal until approximately three and one-half months of age, at which time he first sat up by himself. At this early age it was noted as he sat by himself that he tilted his head to the left. This condition persisted but no medical care was sought for this complaint until the age of four years at which time he was seen in the clinic of another hospital. At this time it was considered advisable that some massage therapy should be administered to the left sterno-cleido-mastoid muscle. The patient was later seen by an orthopedic surgeon in Baltimore who suggested that a leather neck brace be worn. This was done during the next six months. Despite all the treatment and discipline directed toward keeping the head straight, the child persisted in holding the head tilted to the left. In February 1949, the child was seen in both the orthopedic and neurological clinics in Children's Hospital. At this time, a prominent ocular deviation was observed and the child was referred to the Eye Clinic for studies.

When the child was first seen in the Eye Clinic, he had a marked head tilt to the left and the chin was depressed. Vision was normal in each eye, and the refractive error was insignificant. All the internal structures of the eyes were normal. In checking the ocular alignment it became quite apparent that the child had great trouble in depressing his right eye down and in. With the head erect and the eyes in a primary position (that is, straight ahead) the right eye was higher than the left. When the patient moved his eyes to the right, the right eye was not quite as high above the left as it was when the eyes were in the primary position. When the eyes were moved to the left, the right eye was still higher than the left eye than when the eyes were in the primary position. When the patient's head was tilted to the right, the right eye became much higher than the left. When the patient's head was tilted to the left, the eyes were approximately level. With the Worth 4-dot test, the patient demonstrated fusion when allowed to put his head into the position in which he habitually carried it. In all other positions the patient had diplopia.

With the above findings, a diagnosis of right superior oblique paresis associated with ocular torticollis was made.

DISCUSSION

Marshall Parks, M.D.: As has been pointed out, the Ophthalmology Department felt that this case represented a clear-cut right trochlear nerve involvement resulting in paresis of the right superior oblique muscle. The reasons for making this statement are:

1. Objective findings

- a. Head tilt to left, chin depressed, and head turned to the left.
- b. With head erect there is a manifest right hypertropia (right eye higher than the left); as the head is tilted to the right the right hypertropia increases; as the head is tilted to the left there is no hypertropia.
- c. The right hypertropia which is present to a small extent with the head erect and the eyes in the primary position (that is, eyes directed straight ahead) decreases when the eyes are moved to the right, and increases when the eyes are moved to the left.
- d. With the eyes in the left field of gaze, the right hypertropia is greater in the field of depression (that is, eyes directed down and left) and less in the field of elevation (that is, eyes directed up and left).

2. Subjective findings

- a. Diplopia in all fields at all times except when the patient assumes his habitual head position.

The reason that this patient with a right trochlear paresis assumes this habitual head position is that by this means he is provided with binocular single vision. When his head is not in this habitual position, he cannot obtain binocular vision.

The following explains how he obtains binocularity by assuming this particular head position. There are two main actions of the superior oblique; one is depression, the other is intorsion (rolling the eye in toward the nose about an anterior posterior axis). With the eye straight, both of these actions are present. With the eye adducted the action of depression is enhanced and that of intorsion diminished. When the eye is abducted, the action of intorsion is greater than the action of depression. Therefore, the patient will turn his head to the left, allowing the right eye to abduct when looking at an object in front of him. In so doing, the eye is partially taken out of the field of the action of depression.

When the head is level and the eyes in the primary position, there is some depressor action being exerted by the right superior oblique. When looking down there is a good amount of depressor action and when looking up there is little depressor action being exerted by the superior oblique. Consequently, the patient will depress the chin when looking at an object in front of him, allowing the eyes to move up out of the field of depressor action of the superior oblique.

Therefore the combined action of turning the head to the left, allowing the right eye to abduct and depressing the chin, allowing the right eye to elevate, permits the eye to be taken out of the field of depressor action of the superior oblique and therefore eliminates the problem of the right eye being higher than the left. Consequently the eyes are on the same level in this assumed head position.

However, the problem does not end here. As mentioned previously when the eye is abducted the action of intorsion is greatest. Therefore, with the head turned to the left and the right eye abducted, we have the lack of intorsion action of the paretic right superior oblique being most manifest in this position. Because the right eye is not intorted to the degree that it should be, if it were receiving normal tone from the right trochlear nerve, the eye is consequently extorted (that is, rotated outward about an anterior posterior axis). This causes the image seen by the right eye to be tilted and the patient experiences a tilted image diplopia.

We will have to review a fundamental point of physiology to see how the patient solves this problem. As you know, when we tilt our head, there is a reflex emanating from the vestibular apparatus to restore our eyes to where the 12 o'clock-6 o'clock, or vertical, meridian of the cornea is parallel with all vertical lines within our environment. This phenomena is an adjunct in maintaining our proper relations in space. Therefore, if we tilt our head to the left, the right eye is rotated outward (extorted) and the left eye is rotated inward (intorted). In order to rotate the right eye outward, the extorters (that is, the muscles which rotate eyes outward) are stimulated and the intorters are inhibited. So in the normal individual, when the head is tilted to the left, the right superior oblique which is an intorter, receives a message to cease all function. If a paretic muscle wanted to mask itself, what better message could it receive than one directing it to cease all action. Therefore, by tilting the head to the left the torsion action of the right superior oblique is done away with.

From the above description you can follow how the patient can mask the depressor and intorsion actions of the paretic right superior oblique in order that he might maintain binocular single vision.

In conclusion there are two important points in the differential diagnosis of ocular torticollis and non-ocular torticollis.

1. In ocular torticollis the head is always turned in the direction of the head tilt, that is, if the head is tilted to the left, it will also be turned to the left. In non-ocular torticollis, the head is always turned in the direction opposite to that in which the head is tilted. This is due to the anatomy and action of the sterno-cleido-mastoid muscle. If this muscle is contracting or suffering a state of contracture, the patient will turn the head away from and tilt the head toward the effected side.

2. In ocular torticollis due to a paresis of an extraocular muscle, it should be manifest to the examiner upon making the head erect, or tilting the head to the opposite side or observing the eyes move from right to left, that there is a very apparent strabismus. The patient should also experience diplopia except when the head is in the habitually assumed position.

AN UNUSUAL CASE OF PERIRENAL ABSCESS

Case Report No. 159

Harold W. Bischoff, M.D.
William S. Anderson, M.D.

W. J. 46-8649

This five year old colored male entered the Children's Hospital on October 19, 1948 with a chief complaint of abdominal pain and fever of approximately eighteen hours duration. The boy was referred for hospitalization from the Out-Patient-Department where he had been examined and found to have left lower quadrant tenderness and a painful mass in the rectum.

The evening before entry the child began to complain of pain in the lower left quadrant of his abdomen, and, according to the mother, had a fever. The boy was given one half of an aspirin tablet, an enema, and put to bed. The following day the child remained in bed and was lethargic, the abdominal pain persisting throughout the day. From the time of onset until the boy was brought to the hospital, he objected to having his abdomen touched.

There was no history of vomiting, diarrhea, constipation, jaundice, polyuria or polydipsia or joint pains. He had had a head cold and a cough one week before admission; however, this had subsided before the onset of the present illness. The family, birth, developmental, feeding and past histories were non-contributory. The disease history revealed that the child had had diarrhea at 2 weeks of age, pneumonia at 18 months and again at 4 years of age at which time he was hospitalized at Children's Hospital. At 9 months of age the boy developed eczema which persisted until 1947 when he was 4 years of age. During the year prior to this entry he was hospitalized twice because of asthma.

Physical examination revealed an acutely ill, well developed and nourished colored male of stated age lying quietly in bed with the left thigh flexed on the abdomen. The position of this extremity remained unchanged regardless of whether or not the boy lay on either side or on his back.

The posterior cervical lymph nodes were palpable and there were bilateral shotty inguinal nodes. The pupils were round, regular and equal and responded to light. The extra-ocular movements were normal as were the fundi. The faucial tonsils were slightly hypertrophied; however, there was neither tonsillar nor pharyngeal injection. Both tympanic membranes were a pearly-grey and the light reflex was present bilaterally. The heart and lungs were normal. There was a minimum of abdominal movement with respiration. The left lower quadrant showed some rigidity. There was no rebound tenderness. No organs or masses were felt. Mild discomfort to fist percussion over the left costo-vertebral angle was elicited.

At rectal examination, there was good sphincter tone and a mass was encountered on the anterior wall about 6 cm. cephalad to the external anal orifice. This mass measured approximately 3 x 4 x 1 cm. The mass was asymmetrical being somewhat larger on the left of the mid-line and there seemed to be a median sulcus. The mass was of a rather firm, rubbery consistency much the same as that encountered in an adult prostate affected by benign hypertrophy. No other abnormalities were found in the pelvis.

The left thigh remained flexed on the abdomen. Straight-leg raising could be accomplished with either the right or the left leg without any increase in abdominal pain.

The reflexes were active and equal in the upper extremities. In the lower extremities, the knee jerks were hypoactive. The cremasteric reflexes were active and equal bilaterally.

At entry the temperature was 101.8°F rectally. The pulse was 116 per minute, respirations 20 per minute and the blood pressure 102 systolic over 64 diastolic. The evening of the day of admission the temperature rose to 104°F rectally.

On the third hospital day, the rectal mass which upon first examination seemed to be bi-lobar with a median sulcus, was now more diffusely enlarged and there did not seem to be any division between the two sides. By this time there was an upward extension of the mass on the left so that it was most difficult to feel its uppermost limit. The inferior border of the mass also felt as though it was enlarging downward and would eventually overlap its inferior attachment. The mass which originally was resilient and rather firm had now become rather boggy and soft. After each rectal examination, the stool on the glove of the examining finger was normal.

On the sixth hospital day the child was started on procaine penicillin, 300,000 units daily. On the seventh hospital day he appeared more toxic than any time since admission. On this day crystalline penicillin was substituted for the procaine penicillin at a dosage of one million units daily. On the second hospital day, when encouraged, the boy had been able to extend his thigh; however, by the seventh day any attempt to extend the thigh either actively or passively was attended by severe pain which the child localized in the left loin. On the seventh day, some coarse rales were heard at the left lung base posteriorly. By the tenth hospital day the rectal mass was absent. There was some fullness in the left seminal vesicle area. Marked spasm and rigidity of the left loin and back still existed and the child was totally unable to extend the left thigh.

On the seventh day during a moment of quiet and relaxation, the exquisite tenderness of the left loin was found to be replaced by pain of moderate severity. It was possible for the boy to actively extend the thigh to about 140°.

On the twelfth day after admission the left side of the abdomen and left loin, although still somewhat rigid, were soft in comparison with previous examinations. He was able to extend the thigh actively and completely while lying in bed. On the thirteenth hospital day the temperature was 99.2°F. Marked spasm and tenderness were still present in the left flank and the left leg could be extended to about 140°.

Routine urinalyses were all negative. The white-cell count on entry was 17,900 per cu. mm.; 84 per cent of these were polymorphonuclears with 71 per cent segmented, 12 per cent bands and 1 per cent young forms. A sickle-cell preparation taken upon entry was negative. A repeat blood count on the ninth hospital day showed 14,000 white blood cells with 75 per cent polymorphonuclears, 72 per cent being segmented and 3 per cent stab cells. An uncatheterized urine culture showed non-hemolytic *Staphylococcus albus* and hemolytic *Escherichia coli*. A smear of the urethra gently milked down from the prostatic area showed a few gram positive cocci. A blood non-protein nitrogen taken on the fourth hospital day showed 30 mgm. per cent non-protein nitrogen.

Between the third and the thirteenth hospital days, the boy was seen in consultation by a urologist, a general surgeon, a proctologist and an orthopedic surgeon.

During this time it was noted that, although at entry both cremasteric reflexes were active and equal in response, the left cremasteric reflex "was faint and delayed on the left." The consensus of opinion of the consultants was that the patient had either a psoas abscess or a peri-nephric abscess on the left which had dissected down along the course of the left ureter.

A flat plate of the abdomen taken on the second hospital day revealed the psoas muscle and kidney shadows on the right side to be well out-lined. These structures were not well out-lined on the left.

Two days later pyelographic examination of the genito-urinary tract at the end of four minutes revealed dye present in both kidney regions and at the end of ten minutes the calices, pelves, and distal ureters were visualized. The pelves were noted to be of a bifid variety and there was some blunting and dilatation of those on the left. At the end of twenty minutes the left kidney was noted to be slightly displaced to the left and the uretero-pelvic junction was displaced somewhat laterally and inferiorly and further dilatation was noted. The interpretation was that of mild unilateral hydronephrosis on the left with displacement of the left kidney away from the midline.

On the sixteenth hospital day, under nitrous oxide and ether anaesthesia, the left kidney was approached through a left dorsal lumbar incision. Surrounding the left kidney, there was found a fluctuant, well walled-off mass. Upon opening the latter, approximately 90 cubic centimeters of thick,

greenish-yellow pus oozed forth. After thoroughly evacuating the pus, the abscess cavity was seen to lie along the posterior and superior aspect of the kidney, and was quite extensive. There was an abundance of necrotic tissue. Two drains were placed, one at either pole of the kidney and the operative wound closed in routine fashion.



Fig. 1. W.J. Thirty minute intravenous pyelogram film showing the lateral deviation of the left kidney with obstructive manifestations as indicated by the dilatation of the calyces and the pelvis.

Culture of the pus removed at operation revealed the presence of hemolytic *Staphylococcus aureus*. Microscopic examination of the necrotic tissue removed during operation showed numerous large polygonal cells with rather large oval nuclei. The typical architecture of the tissue could not be identified; however, the cells closely resembled those seen in the adrenal gland. Marked leucocytic infiltration and evidence of hemorrhage was observed.

Within 36 hours after operation the child's temperature dropped to normal. Crystalline penicillin, 1,000,000 units daily, was continued for five days after operation. The boy was discharged in good condition on his thirty-second hospital day.

DISCUSSION

According to Callander⁽¹⁾ perinephric abscess or abscess in the lumbar extraperitoneal fatty areolar tissue as a secondary process rarely occurs following primary pathologic change in the kidney parenchyma, pelvis or ureter. Whether the perinephric abscess is of renal or non-renal origin is of little clinical importance and the location of a pre-existing infection, or focus of infection actually a point of academic interest.

Considering for the moment that the perirenal fascia (Gerota's Fascia) extends downward over the ureter and behind the bladder and is continuous with the fascia covering the seminal vesicles and prostate^(2a), it is not difficult to imagine how an infection primarily localized in the kidney bed could point downward and present in the region of the prostate and left seminal vesicle.

It was this downward extension and the contiguity of the infected peri-ureteral tissues with the ilio-inguinal nerve which apparently gave rise to the lack of response of the left cremasteric reflex as the process progressed. Furthermore, since there was resolution of the rectal mass following the institution of penicillin therapy, it is probably safe to assume that the infection began above and proceeded downward rather than the reverse sequence of events.

The onset of the clinical picture in perirenal abscess may be acute or chronic^(2b). Certainly, the case under consideration, with a clinical history of eighteen hours duration, should be considered as acute in onset. However, considering the "well walled-off mass" found at operation, this infection may well have been present for some time before it became manifest by overt clinical signs. Another possibility is, of course, that the penicillin contributed to the walling-off process. Once the abscess capsule had been formed, it was impossible for the penicillin to penetrate the inflammatory tissue barrier to sterilize the infectious material within the abscess cavity. In the case reported by Ockuly, Barnhart and Egbert⁽³⁾, if theirs were truly a perinephric abscess, penicillin treatment must have been begun before the formation of such a capsule. Helmholtz⁽⁴⁾ states that the infecting organism is practically always the staphylococcus. Bergman and Simon⁽⁵⁾ found this to be true in their recent review of twenty-two cases of perinephric abscess. That this was true in the present case was borne out by the recovery of hemolytic *Staphylococcus aureus* from the pus obtained at operation. There was no previous history of pyoderma, otitis or osteomye-

litis so that it becomes difficult to localize the portal of entry of the organism. One point in the history is perhaps helpful in supposing a portal of entry. The mother stated that the boy had had a head-cold and a cough one week before hospital entry. Since the child had not been seen by a physician at that time, it cannot be stated whether or not he had had a bacterial (Staphylococcal?) infection of his naso-pharynx and/or middle ear with subsequent blood stream metastasis.

Another point of anatomical interest is the finding of cells, presumably of adrenal origin, in the pathologic specimens of necrotic tissue obtained at operation. The adrenal glands are enclosed in a special compartment of the perinephric fascia of Gerota^(3c). Here again we might reason from our knowledge of the anatomy of the region that an accumulation of pus of sufficient magnitude and under enough tension to dissect downward along the fascial layers surrounding the ureter, could, by the same token, also ascend, destroying the fascial barrier and involve the compartment occupied by the adrenal gland.

SUMMARY

1. An unusual case of perirenal abscess in a five year old colored male is presented.
2. No definite, previously existing, focus of infection could be delineated as the cause of the perirenal abscess.
3. The abscess presented in the region of the left seminal vesicle.
4. The anatomy of the region is briefly reviewed and a correlation between those considerations and the clinical picture is suggested.

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LEUKOPENIA AND DRUG FEVER IN A JUVENILE HYPERTHYROID TREATED WITH PROPYLTHIOURACIL

Case Report No. 160

Hugh J. Davis, M.D.
Frank J. Murphy, M.D.

A review of the literature reveals little mention of the use of propylthiouracil in the pediatric age group. One reason for this may well be the infrequency of hyperthyroidism in the young patient. During the past thirteen years there were only six cases of this disease seen at the Children's Hospital. With this in mind the authors feel that a case report of a pediatric patient treated with propylthiouracil may be worthwhile.

The following is a case presentation of a 4 year old child admitted with classical symptoms of hyperthyroidism who developed leukopenia and a drug fever while on propylthiouracil therapy.

M. W., a four year old white female, was admitted to the hospital on September 19, 1947 because of exophthalmos and nervousness.

The child had been in apparent good health except for her failure to gain weight in spite of an adequate food intake for the previous six months. Four weeks prior to entry a gradual progressive bulging of her eyes was first noted, associated with an increasing degree of nervousness characterized by constant chattering and agitated movements. These symptoms had been apparently masked at the onset by a generalized fine rash which was distributed mainly over her extremities but also involved her trunk and face. At that time, a diagnosis of measles had been entertained; however, the rash was subsequently noted to fade and reappear several times. About one week before admission the patient complained of a fullness in her throat but exhibited no difficulty in breathing or swallowing. There was no history of any febrile period prior to entry.

The past and family histories were non-contributory to the present illness.

Physical examination revealed a well developed and fairly well nourished child who was very hyperactive with a moderate exophthalmos but not otherwise ill. The temperature on admission was 99.6°F, pulse 148 and blood pressure 138 systolic and 60 diastolic. Her weight was 38 pounds. There was a discrete generalized macular rash, more marked over the lower extremities but also involving the trunk. Moderate exophthalmos was present. The pharynx was slightly injected. There was a non tender, movable walnut sized mass present in the thyroid area. The liver was palpable 2 cm. below the right costal margin.

Examination of the blood showed 13 gm.% of hemoglobin, 4,000,000 erythrocytes, 8,200 leukocytes with a differential of 65% neutrophils and 38% lymphocytes. Urinalysis was negative. The basal metabolic rate on three successive occasions was plus 73, plus 52 and plus 72. The blood cholesterol was 110 mg.%.

On the sixth hospital day the patient was started on 150 mg. of propylthiouracil daily. The course was uneventful until seven days later when the temperature rose and the leukocyte count fell to 2,000 with a differential of 46% neutrophils and 42% lymphocytes. Propylthiouracil was then discontinued and during the succeeding five days the temperature ranged between 100–105°F and was associated with an excrescence of the macular rash over the entire body. Examination revealed nothing more than an injection of the pharynx. Intramuscular penicillin was administered as a prophylactic measure during this period. With the decline in temperature, the rash faded and the leukocyte count increased to 6,000 with a differential of 20% neutrophils.

On the twentieth day, propylthiouracil was restarted with a reduced dosage of 50 mg. daily. Forty-eight hours later the fever returned, the rash reappeared and the leukocyte count dropped to 4,600 with 13% neutrophils. The drug was again stopped and penicillin re-employed followed by a subsidence of the fever and rash. However, on the thirty-eighth day her leukocyte count was 3,800 with the neutrophils down to 2%. The patient was given parenteral liver and pyridoxine with a gradual return to a normal leukocyte picture.

The child was subsequently prepared for surgery and a subtotal thyroidectomy was successfully performed.

Follow-up examination over an 18 month period shows the patient to be in a state of well being with a normal hemogram, an increase in weight and a basal metabolic rate within normal limits.

DISCUSSION

A review of the literature indicates that toxic symptoms are manifested with the use of thiouracil in approximately 15% of patients^(1, 2, 3) and in about 1.2%^(3, 4) with propylthiouracil. These undesirable reactions include fever, leukopenia, drug rash and rarely agranulocytosis. There is a paucity of pediatric cases treated with the latter drug in the literature and while one instance does not indicate the frequency of toxicity of the pediatric age group, it does serve to emphasize the point that these drugs must be given with caution. It is hoped that wider clinical experience with propylthiouracil will permit as favorable an outlook in the treatment of hyperthyroidism in children as it apparently offers in the adult patient.

SUMMARY

A case of leukopenia and drug fever in a 4 year old hyperthyroid patient treated with propylthiouracil is reported. The remission of symptoms on withdrawal of the drug followed by the reappearance of fever and leukopenia after propylthiouracil was again used is noted.

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CLINICO-PATHOLOGICAL CONFERENCE

Directed by: E. Clarence Rice, M.D.

Assisted by: John E. Cassidy, M.D.

D. Joseph Judge, M.D.

Edwin Vaden, M.D.

By Invitation: Arthur Sims, M.D.

Dr. Judge:

T. B. 48-1268

T. B., a 6 month old white male infant, was admitted to the hospital during the month of February with the chief complaint of a generalized skin rash of approximately eight hours duration.

During the two weeks prior to admission the infant had a slight cold. Constipation was the only other symptom. On the day before admission, the child ate well and seemed normal. About eight hours before coming to the hospital the mother noticed that he had a rather generalized, sparse, light red rash. Shortly afterwards the child developed a fever, vomited and the rash increased in extent and number of lesions. In about six hours the patient's skin had become blotchy and bluish and shortly afterwards he became stuporous and began to breath noisily and rapidly. No other symptoms or signs were noted.

The child was born by caesarian section. Development had been normal. A circumcision had been performed two weeks prior to admission and the site had become swollen but apparently not infected. The past and family histories did not contribute any pertinent information.

Physical examination disclosed a well developed and nourished white male child apparently in circulatory collapse. The skin had a bluish-white pallor and there were multiple small and large petechiae over the entire body. The pulse was 140, respirations 24, the temperature 103.2°F. The periorbital tissues appeared edematous. The pharynx was injected. The extremities were flaccid. There were no other positive physical findings.

The child was given glucose in physiologic salt solution, plasma and 30,000 units of penicillin by bone marrow injection. Five hundred thousand units of penicillin and 10 ml. of adrenal cortical extract were immediately given intramuscularly. Oxygen was administered continuously. Blood culture was taken which later revealed no growth.

In spite of all therapeutic and supportive measures the child expired one hour and 45 minutes after admission and approximately 10 hours after the onset of symptoms.

DISCUSSION

Arthur Sims, M.D.: We are presented here with the case of a 6 months old infant who suddenly became ill with fever, vomiting and a petechial

rash, and who rapidly progressed to a fatal outcome within 10 hours. There are several diagnostic possibilities which must be considered.

Because of the nature of the rash one must consider the possibility of purpura hemorrhagica or Werlhof's disease. This disease is usually classified into three categories, the rarest of which, fortunately, is the acute fulminating type. In this condition the onset is usually abrupt with a petechial rash which may rapidly become extensive and produce death in a short time. It may or may not be accompanied by fever so that the presence of fever in this particular case does not lend argument against this diagnosis. However, in acute purpura hemorrhagica one would expect to find bleeding from the mucous membranes and the body orifices, and in fact this feature is often the more prominent one. I believe on this basis alone one may fairly well eliminate this possibility in this case. There is perhaps an additional argument against this diagnosis. If we can assume, and I believe we can, that the usual routine blood studies were performed prior to circumcision done on this child two weeks before the onset of the present illness, one would expect the chances to be good of finding an abnormal bleeding time then. I do not suppose that there was time for a bleeding time, platelet count, and clot retraction time to have been done in our case.

Dr. Judge: No other laboratory work was performed.

Dr. Sims: The second diagnostic possibility that I think should be considered is purpura fulminans, a condition not to be confused with acute fulminating purpura hemorrhagica. In purpura fulminans the onset is characteristically abrupt with a petechial rash and a high fever, and thereafter rapid progression with further hemorrhage into the skin, circulatory collapse and death within a matter of several days. This condition is similar in all respects to that seen in acute overwhelming septicemia with purpura except that in the former condition positive bacteriological evidence is lacking, the blood cultures and post mortem bacterial examinations being negative for infectious origin. I do not feel that with the evidence presented in our case we can dismiss this diagnosis.

The third condition that I would like to bring up for consideration is that of a very severe case of acute hemorrhagic measles. In this condition there is an early association with the hemolytic streptococcus with subsequent bleeding into the skin lesions and frequently early circulatory collapse and death. However, I feel that this diagnosis can be readily eliminated on the basis that our patient failed to show the usual prodromal symptoms of measles. I do not feel that the "cold" symptoms our patient had during the two weeks prior to the onset of the present illness can be interpreted as prodromata of measles. One would certainly expect to find rather marked naso-pharyngeal and bronchial symptoms in addition to the presence of fever on the day before the rash appeared. Our history states that on the

day before admission the child was normal in all respects. Further evidence against measles if such is needed might be looked for in the fact that we are dealing with a 6 months old infant who, if the mother falls into the 90-95% of adults who have had measles, would still possess some degree, minor as it may be, of prenatally acquired immunity.

The next condition which I would like to mention is that of Rickettsial disease. It is perhaps possible that Rocky Mountain spotted fever might present a fulminating course; however, I have never heard of a case presenting so rapid a progression of symptoms to death as we are confronted with here. Usually in Rocky Mountain spotted fever the rash appears some 2 to 5 days after the onset of symptoms. In addition, the season of the year and the age of the child speak highly against this diagnosis. I feel that we can readily eliminate it as a possibility in this case.

Such causes of purpura as leukemia, aplastic anemia, drug sensitivity, allergic manifestations and acute bacterial endocarditis can be quickly dismissed by virtue of the overwhelming nature of our case as well as the lack of sufficient evidence to consider them further.

We are now left with the possibility of an acute overwhelming septicemia with purpuric manifestations. The vast majority of these cases are caused by the Meningococcus and so I will refer to this as meningococcemia; however, it must be made clear that in a small percentage of cases, other organisms may be at fault. Perhaps *H. influenzae* deserves special mention among this category. When a child presents himself with fever and a petechial rash the best diagnosis is that of meningococcemia unless otherwise proven. In this condition there is usually an abrupt onset following a history of a common cold which may have been present for several days or several weeks. The onset is then characterized by fever, petechiae, nausea and vomiting and there may or may not be convulsions, joint manifestations, and signs of central nervous system involvement. This of course fits well with our case on hand. The periorbital edema noted in this child is very frequently seen in cases of meningococcemia. In a certain percentage of cases meningococcemia proceeds with rapid extension of skin hemorrhages leading to circulatory collapse and death within a matter of hours. When this occurs the name Waterhouse-Friederichsen syndrome is applied. This syndrome was formerly considered to be due to adrenal hemorrhage; however, more recent investigation has tended to show that the cause of shock and death is probably due to the overwhelming toxemia with secondary capillary damage and increased capillary permeability which permits of hemorrhage into various organs of the body, one of which may or may not be the adrenal gland. The diagnosis must look to the laboratory for definite confirmation. First of all a leucocyte count and differential would be helpful. If it had been done I would expect to find a leukocytosis with the total

white count being somewhat over 18,000 and more likely in the vicinity of 25,000. Even more characteristic would be the differential count in which one would probably find a shift to the left with perhaps over 90% of the cells falling into the polymorphonuclear series. The second laboratory procedure which I think of importance is probably the most rapid and most practical method of making a diagnosis; this consists of making a smear of a skin lesion, preferably a fresh one, and of staining it with Wrights and Grams stain. In the vast majority of cases one would find both pus cells and gram negative diplococci. Another laboratory method of value is the blood culture. In this case the fact that the one blood culture was negative I do not feel can, in any way, eliminate this diagnosis. The meningococcus is very fastidious in its growth characteristics. I presume that the growth was cultured under CO₂ tension.

The other laboratory procedure which might be of some diagnostic value would be that of agglutination blood studies.

In conclusion I feel that we are dealing here with a case of acute fulminating septicemia probably due to the meningococcus. However, if there is complete lack of bacteriological evidence I suppose that for terminological reasons I would then have to call it purpura fulminans, cause undetermined.

PATHOLOGICAL DISCUSSION

John E. Cassidy, M.D.: Postmortem examination confirmed the clinical impression in this case, namely that of an overwhelming septicemia due to the meningococcus with bilateral adrenal gland hemorrhage. Both adrenal glands were enlarged to twice normal size due to the presence of extensive hemorrhage. The skin and serous membranes of the body cavities revealed a generalized petechial rash. The leptomeninges grossly were normal but microscopically there was a slight scattered polymorphonuclear cell infiltration. Bacteriologic studies revealed the presence of *Neisseria intracellularis* in the adrenal glands and in the cisternal fluid.

This picture comprises the syndrome known as Waterhouse-Friederichsen syndrome. Dr. Sims has brought out the important points in the diagnosis and the differential diagnosis. I would like to stress again the value of examination of the petechiae by smears and culture in obtaining information as to the organism causing the disease. It is a relatively simple procedure and the organisms can be recovered in a high percentage of cases.

The exact cause of the rapid progression of symptoms and early death in these patients probably results from a combination of the overwhelming toxemia, capillary damage, and adrenal insufficiency as a result of the extensive hemorrhage into the glands.

There are cases reported in the literature of recovery from Waterhouse-Friederichsen syndrome. Some doubt arises in ones mind concerning these

especially if one holds that adrenal gland hemorrhage must be present before this name can be applied. We have never seen a recovery in this hospital. However, constant efforts toward earliest possible diagnosis and institution of early vigorous treatment may yield beneficent results.

It is of interest to note that there were very early inflammatory changes in the lepto-meninges. This has been our experience in the past. Most of these patients die before a full blown meningitis can develop.

We wish to thank Dr. Sims for being with us and to congratulate him for his fine discussion.

SHOULD VITAMIN D BE GIVEN ONLY TO INFANTS?

VITAMIN D has been so successful in preventing rickets during infancy that there has been little emphasis on continuing its use after the second year.

But now a careful histologic study has been made which reveals a startlingly high incidence of rickets in children 2 to 14 years old. Follis, Jackson, Eliot, and Park* report that postmortem examination of 230 children of this age group showed the total prevalence of rickets to be 46.5%.

Rachitic changes were present as late as the fourteenth year, and the incidence was higher among children dying from acute disease than in those dying of chronic disease.

The authors conclude, "We doubt if slight degrees of rickets, such as we found in many of our children, interfere with health and development, but our studies as a whole afford reason to prolong administration of vitamin D to the age limit of our study, the fourteenth year, and especially indicate the necessity to suspect and to take the necessary measures to guard against rickets in sick children."

*R. H. Follis, D. Jackson, M. M. Eliot, and E. A. Park: Prevalence of rickets in children between two and fourteen years of age, *Am. J. Dis. Child.* 66:1-11, July 1943.

MEAD'S Oleum Percomorphum With Other Fish-Liver Oils and Viosterol is a potent source of vitamins A and D, which is well taken by older children because it can be given in small dosage or capsule form. This ease of administration favors continued year-round use, including periods of illness.

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